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Subcutaneous Sacrococcygeal Myxopapillary Ependymoma

Inci Ilhan, MD,[1]* Semha Berberoglu, MD,[1] Lale Kutluay, MD,[2] and Hasim Ata Maden, MD[3]

We report an 8-year-old boy with a primary subcutaneous sacrococcygeal ependymoma, a rare tumor that is thought to arise in embryologic rests. The lesion was completely removed in our patient, who has been followed without recurrence for 20 months. Our experience, together with that of the other 15 cases in the world literature, supports surgical excision as the mainstay of treatment. Med. Pediatr. Oncol. 30:81–84, 1998. © 1998 Wiley-Liss, Inc.

Key words: tumors; childhood cancer; extra-axial ependymoma

INTRODUCTION

Myxopapillary ependymomas are unusual but distinct lesions that typically occur in the distal segment of the spinal cord [1]. In rare instances, myxopapillary ependymomas may occur as a primary lesion in the postsacral soft tissues [2], and about 50 cases of ependymoma in extraspinal locations have been reported in the literature [3–5].

We report on the clinical and pathologic findings of an extradural ependymoma found in a child who was treated with surgery and subsequently followed with no additional treatment and without recurrence 20 months after surgery.

CASE REPORT

An 8-year-old boy was admitted to the hospital in June 1995 because of a mass in the sacrococcygeal area that had gradually enlarged for 3 months. There was no history of inflammation or drainage. Neurologic and physical examinations were normal except for the presence of a subcutaneous 3 cm mass over the tip of the coccyx. Rectal examination showed no mass. Routine preoperative laboratory studies, chest X-ray films, thoracic computed tomography (CT), lumbosacral spine films, and magnetic resonance imaging (MRI) were normal. Spina bifida was not detected. A preoperative sacrococcygeal CT (Fig. 1) showed that the tumor was confined primarily to the subcutaneous tissues with extension down to the sacrum but not beyond. It was removed en bloc with the coccyx.

The gross specimen consisted of an encapsulated nodular tumor measuring 5 × 3 × 2 cm with a small fragment of bone 0.8 cm in diameter attached to it. Examination of a cut section revealed that the lesion was solid, white in color, and showed a lobular growth pattern. The whole specimen was routinely processed with the fragment of bone which was decalcified before examination.

Microscopic sections revealed a tumor consisting of micropapillary projections with a central vascular core and covered by one or two layers of uniform cuboidal cells. The papillary structures were embedded in a stroma showing focal hyalinization and myxoid degeneration (Fig. 2). There was no mitotic activity, nuclear pleomorphism, or atypia. The tumor was surrounded by a thin fibrous capsule. The bone was intact and showed no signs of invasion.

Immunohistochemical study was performed with a monoclonal antibody directed against glial fibrillary acidic protein (GFAP; 1:1,000, Dako, Carpinteria, CA) and S-100 protein (Brotech Solutions, prediluted) using the peroxidase-antiperoxidase method. The tumor cells were strongly stained with anti-GFAP, but were negative for S-100 protein.

Flow cytometric study was carried out on a paraffin-embedded tissue block. It revealed diploid DNA content (DNA index 1.0) with an S-phase of 8%. Normal human lymphocytes are used as the diploid control. In our laboratory, the upper limit of low proliferative activity is 7%, and 8% S-phase is considered low to intermediate proliferative activity.

The child recovered uneventfully and is doing well, without recurrence, 20 months following resection.

[1]Department of Pediatric Oncology, Ankara Oncology Hospital, Ankara, Turkey.

[2]Department of Pathology, General Pathology Center, Ankara, Turkey.

[3]Department of Pediatric Surgery, Dr. Sami Ulus Children's Hospital, Ankara, Turkey.

*Correspondence to: Inci Ilhan, 37 Sokak 13/8, Bahcelievler-Ankara 06500, Turkey.

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Fig. 1. Pretreatment axial sacrococcygeal CT image. A mass involving the fat tissue in the midline posterior of the rectum is seen. The mass is well defined and isodense with the gluteal muscles.

DISCUSSION

Ependymomas are the most common tumors of glial origin of the spinal cord. They account for 50% of the neoplasms of the filum terminale [6]. In this location, they are frequently of the papillary or myxopapillary type. The sacrococcygeal myxopapillary ependymoma of soft tissue is a rare tumor in children and young adults [5,7–11].

Helwig and Stern [12] discussed four situations of involvement outside the central nervous system: 1) metastasis or direct extension following surgical excision of a primary tumor from the central nervous system, 2) direct extension to the soft tissues of the sacrococcygeal area from a primary ependymoma of the lower spinal cord, cauda equina, or filum terminale, 3) occurrence as a primary tumor of the skin, and 4) subcutaneous tissue of the sacrococcygeal area without demonstrable connection with the spinal cord. This case would fall into the fourth category.

Most of the subcutaneous sacrococcygeal ependymomas have been clinically diagnosed as pilonidal cysts (postsacral), teratomas, or chordomas (both of them are presacral) [12]. However, immunohistochemical study can rule out these diagnoses, since a constant positivity is observed with anti-GFAP antibody in almost all cases of ependymomas. This positivity is often described with a less constant staining with anti-S-100 protein, anti-vimentin, and anti-keratin antibodies [13].

The origin of ependymal neoplasm is unclear. It is generally agreed that subcutaneous sacrococcygeal epen-

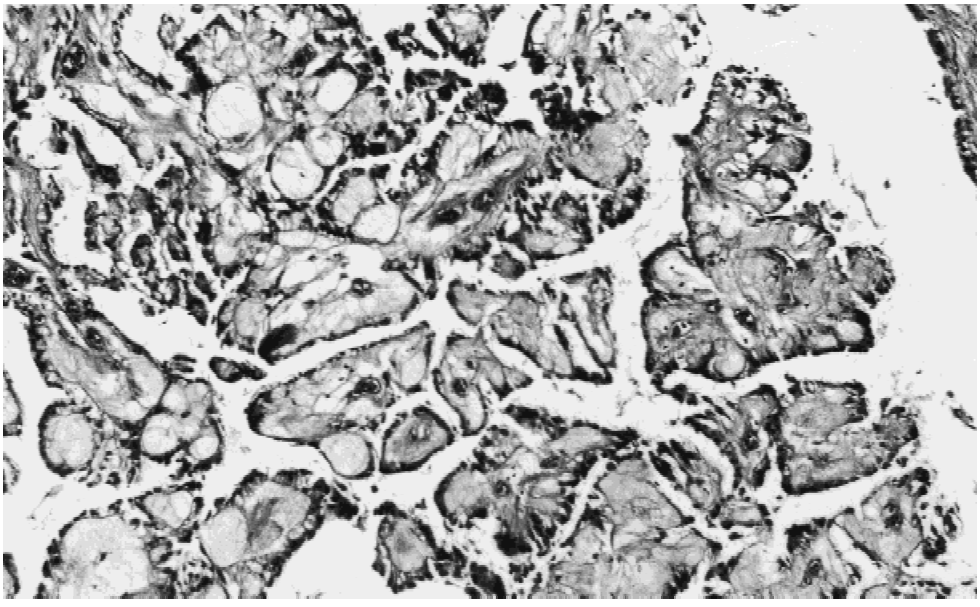

Fig. 2. Sacrococcygeal ependymoma. The tumor is composed of papillary projections that line irregular cystic spaces (hematoxylin-eosin, ×100).

TABLE I. Reported Cases of Extradural Ependymomas of Dorsal Sacrococcygeal Region in 15 Children

Reference	Patient's age/sex at 1st surgery	Duration of symptoms prior to operation	Surgical procedure related to this lesion	Length of follow-up	Local recurrence	Sites of metastases
[20]	16 years/M	9 years	Yes × 3	Not known	Yes	Inguinal lymph nodes, pelvis
[1]	3 years/M[a]	Not known	Yes	Not known	Yes	Not known
	8 years/F	Not known	Yes	5.5 years	No	No
	16 years/F	Not known	Yes	2 years	No	No
[16]	4 years/M	<1 year	Yes × 1	20 years	No	Inguinal lymph nodes
[4]	4 years/F	4 weeks	Yes × 1	10 years	No	No
[11]	11 years/F	10 months	Yes × 1	7 months	No	No
[5]	9 months/F	Since birth	Yes × 1	Not known	No	No
	2 months/F	Since birth	Yes × 1	6 years	No	No
	4 months/F	Since birth	Yes × 1	42 months	No	No
	11 months/M	Since birth	Yes × 1	9 months	No	No
	26 months/M	Since birth	Yes × 1	5 months	No	No
[10]	5 days/M	Since birth	Yes × 1	Not known	No	No
[9]	14 years/F	6 months	Yes × 1	2 years	No	No
[8]	18 months/F	Since birth	Yes × 2	Not known	Yes	Inguinal lymph nodes

[a]This was the only patient to receive radiotherapy, and the dose is not known.

dymomas arise from embryologic rests such as extraneural extensions or remnants of the filum terminale [14]. A remnant of the caudal portion of the neural tube remains beneath the skin over the end of the coccyx as an ependyma-lined cavity known as the coccygeal medullary vestige and is the source of ependymal neoplasms [2–5]. Spina bifida occulta occurs in 20–30% of patients with extradural ependymomas [6].

Unfortunately, there is no correlation between the histologic appearance and subsequent behavior of these tumors [8]. They metastasize to regional lymph nodes, lung, bones, and liver in up to 20% of cases after a prolonged disease-free interval, sometimes up to 10 years [6,15,16]. Most authors believe that operative intervention opens up vascular and lymphatic spaces to the tumor cells and thereby promotes distant metastases [6]. The interval between detection and resection of the primary lesion and development of distant metastases may be long, showing that the sacrococcygeal myxopapillary ependymoma is a malignant tumor of low grade [17].

In the literature, the determination of DNA content was performed in a young adult patient by Sapi et al. [18]. The tumor proved to be aneuploid with low proliferative capacity in spite of absent histologic signs of malignancy. The DNA content in our case was diploid. No reliable inference about the biologic behavior of these tumors based on DNA content can be drawn from these two cases.

Adequate treatment of subcutaneous sacrococcygeal myxopapillary ependymoma consists of wide excision [6,19]. Complete removal is technically feasible for most postsacral tumors; however, for presacral lesions, the very invasive nature of the tumor may demand a combined procedure such as a neurosurgical and pelvic approach [19]. The influence of radiation therapy on the metastatic potential of these neoplasms has also been widely discussed. There is, however, a clear lack of data with which to address this question and no definite dose-response relationship could be detected in the few studies done [6,12]. Radiotherapy is usually recommended for residual or inoperable tumors [7]. There also are no data about the usefulness of chemotherapy, so its role in these tumors remains uncertain. In our case, we performed wide complete excision, and follow-up 20 months later indicated no recurrence. Even though the postoperative survival period of our patient is short, some reports—especially those about this lesion in children and young adults—show that such patients can exhibit long-term event-free survival after surgery without additional treatment [5,8–11].

After the first report by Hendren and Hardin [20] of this lesion in children, 15 additional cases appeared in the literature. The relevant features of previously published pediatric cases are summarized in Table I. Among them, local recurrence of tumor developed in three patients, but only one required postoperative irradiation.

In conclusion, sacrococcygeal extradural ependymomas must be considered in the differential diagnosis of presacral or postsacral mass lesion, and complete surgical removal should be attempted in all cases. We are aware that, although the postoperative survival period can be long, these tumors tend to metastasize more frequently than the intradural variety, and a follow-up of only 20 months is not long enough to decide that our patient has been completely cured.

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